INTRODUCTION

- Erythromelalgia (EM) is a rare and debilitating syndrome presenting with burning pain, erythema, and swelling in the extremities.
- Potential mechanisms for EM include arteriovenous shunting, small fiber neuropathy, or inherited via mutations in voltage gated sodium channels.
- Treatment can be challenging and multiple medications trials are often needed to identify a strategy that reduces symptoms without significant side effects.
- We present the case of an adolescent male presenting with EM discovered during a rheumatological evaluation and treatment for juvenile idiopathic arthritis (JIA).

CASE HISTORY

History of present illness: The patient is an 18 year old male who presented with complaints of pain in the lumbar spine and swelling of the toes on the right foot. Initially diagnosed with enthesis and juvenile idiopathic arthritis, he was placed on Entrel and followed by rheumatology. Concurrent anxiety was observed and the patient was started on sertraline and clonazepam PRN. Over the next year, the patient report painful burning and swelling in the hands and feet. Reynaud’s was considered but the patient reported relief of his symptoms following submersion of the painful extremities in cold water. EM was suspected and the patient was tested for and found to be negative for the SCN9A mutation. He was referred to our pain clinic for further management.

Treatment Course:
- Antidepressants: presented on sertraline and clonazepam with good anxiety control but little improvement in EM symptoms
- Aspirin and oral amitriptyline: previously trialed and ineffective
- Gabapentin – effective initially but stopped due to intolerable dizziness.
- Pregabalin – ineffective.
- Mexiltiline – stopped due to dizziness and nausea.
- Topical Cream – Gabapentin 6%, lidocaine 2.5%, prilocaine 2.5%, isopropylate 1%, and meloxicam 1% (applied once nightly).
- This therapy improved our patient’s hand symptoms and moderately improved his feet symptoms without side effects

DISCUSSION

Erythromelalgia Epidemiology
- Female 2.2:1 male, average age 56, w/ range 5-91
- Occurs in idiopathic or inherited fashion via a mutation in the SCN9A gene (encodes Na-channel protein subunit)
- In a study of 32 children with the Mayo Clinic
- Female 69%, male 31%, mean age 14, incidence 1.3 in 100,000
- Common presentation: intermittent symptoms, all had symptoms in the feet, half with symptoms in the hands; skin described as having a purple hue, cool, and painful
- Common treatment regimens in children: ASA, NSAIDs, antidepressants, antihistamines, vasodilators, and beta blockers

IMAGES

Figure A. Clinical signs of erythromelalgia flare, including erythema, as demonstrated by the hand on the left. The hand on the right, in contrast, is normal.

Figure B. Clinical signs of erythromelalgia flare on the foot of the patient.

Figure C. Additional demonstration of signs of erythromelalgia on bilateral hands including patchy erythema and swelling.

CONCLUSION

- EM is rare and can be challenging to treat as multiple therapeutic trials may be necessary.
- Topical agents have been reported to be effective in treating EM.
- We struggled to manage our patient with systemic agents and turned to a local compounding pharmacy to develop an analgesic cream.
- More investigation is needed, but practitioners caring for patients with EM may want to consider topical agents as a treatment option.

REFERENCES


Table 1. Potential treatment approaches used in managing erythromelalgia

<table>
<thead>
<tr>
<th>Treatment</th>
<th>Potential Side Effects</th>
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<tbody>
<tr>
<td>Aspirin</td>
<td>GI complaints, nausea, headache, hypotension, drowsiness</td>
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<td>TCAs</td>
<td>GI complaints, nausea, headache, hypotension, drowsiness</td>
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<td>Gabapentin</td>
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<td>Magneum</td>
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<td>Ketamine</td>
<td>GI complaints, nausea, headache, hypotension, drowsiness</td>
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